216 A UK survey on the assessment and management of CF-associated liver disease

D. Honeybourne[1], E.F. Nash[1], P.N. Newsome[2], J.L. Whitehouse[1]. [1]*Heartlands Hospital, West Midlands Adult CF Centre, Birmingham, United Kingdom;* [2]*University Hospital Birmingham, Liver Unit, Birmingham, United Kingdom*

This survey was designed to assess the amount of variation around the UK in the investigation and management of CF related liver disease (CFLRD). Questionnaires were devised by a group of adult and paediatric CF physicians and also hepatologists and were sent to all major CF centres in the UK. 16 adult and 24 paediatric centres replied. The number of new CFLD cases ranged from 0–5 per year per centre. No centre carried out routine diagnostic liver biopsy and most centres did not use a severity scoring system. Non-invasive imaging other than ultrasound was mentioned by only 5 centres. In established CFLD, all centres monitored platelets, prothrombin time and albumin. Ultrasound monitoring varied between 6 monthly, annually or ad hoc. There was wide variation in screening for varices. Only 16 centres ran a joint clinic with a hepatologist but 38 centres had access to a hepatologist with a special interest in CFLD. Nearly all centres used lifelong URSO treatment for patients with, doses of 10–30 mg/kg/day but only 5 centres used taurine. All adult centres advocated primary prophylaxis for grade 2+ varices and the majority preferred banding to beta blockade. Most centres considered triggers for referral for liver transplant to be one or more of: ascites, bleeding varices and impaired synthetic function but there was less consensus on proximal muscle wasting. 15 out of 40 centres felt that they did not have an adequate forum to discuss liver transplant issues. In summary, our survey showed wide variation in the assessment and management of CFLD in the UK with some differences compared to recent European guidance [1].

Reference(s)

[1] Debray D et al, 2011, J Cystic Fibrosis; 10: S29–36.

217 Use of liver ultrasound in assessment of cystic fibrosis liver disease in children

P. Shukla[1], E. Twomey[2], J. Wagener[1,3], D. Slattery[1]. [1]*Children's University Hospital, Respiratory Department, Dublin, Ireland;* [2]*Children's University Hospital, Radiology Department, Dublin, Ireland;* [3]*University of Colorado Denver, Department of Pediatrics, Aurora, United States*

Objectives: While pulmonary disease remains the main cause of morbidity and mortality in cystic fibrosis patients, Cystic Fibrosis Liver Disease is assuming increased importance as patients with CF live longer. Early identification is difficult to diagnose clinically or biochemically. We aim to assess effectiveness of liver ultrasound in CF patients in identifying progression from normal scan towards CFLD.

Methods: Retrospective review of sequential liver ultrasounds over 10 years were used to identify trends in CFLD progression. Comparisons of serial liver scans in individual patients were quantified based on radiological reports from a tertiary children's hospital. Radiological reports were graded on basis of progression towards liver disease.

Conclusions: 101 CF patients were identified over ten years. 59 were male. 27 were excluded for having undergone one or less scans. 401 scans were reviewed in total; 94 [23%] were abnormal. 54 [13%] scans in 29 patients showed fatty infiltration. 18 [4%] scans in 10 patients demonstrated increased echogenicity with non-specific changes. 22 [5%] studies in 6 patients had liver cirrhosis. Liver disease progression was assessed by comparison of subsequent scans in individual patients. 258 [81%] of 320 comparisons showed no change. 29 [9%] scans subsequently showed progression of disease. Interestingly, 33 [10%] showed reversion towards normality. It is unclear whether this is related to interoperator variability, a characteristic of CFLD, or due to a combination of both.

While independently, ultrasound has diagnostic limitations, it remains a useful tool in identifying early and late CFLD when used in conjunction with clinical exam.

218 Liver elasticity in CF associated liver disease

I. Ciuca[1], L. Pop[1], Z. Popa[2], A. Deleanu[3], S. Turcu[4], B. Guta-Almajan[5], L. Tamas[6], I. Popa[7]. [1]*University of Medicine and Pharmacy Victor Babes, Pediatric II Department, Timisoara, Romania;* [2]*Cystic Fibrosis Centre, Timisoara, Romania;* [3]*University of Medicine and Pharmacy Victor Babes, Gastroenterology Department, Timisoara, Romania;* [4]*Evelina Children's Hospital, London, United Kingdom;* [5]*University of Politehnica, Sport and Physical Education, Timisoara, Romania;* [6]*University of Medicine and Pharmacy Victor Babes, Biochemistry, Timisoara, Romania;* [7]*University of Medicine and Pharmacy Victor Babes, Timisoara, Romania*

Cystic fibrosis liver disease (CFLD) is the second cause of death in cystic fibrosis (CF) after pulmonary disease, with an increasing prevalence. The main pathogenic lesion of cirrhosis is liver fibrosis who's early detection could predict the following outcome. Elastography seems to be a useful tool in liver fibrosis assessment.

Objectives: Evaluate the utility of the transient elastography in addition to ultrasound for CFLD early diagnosis and monitoring in our CF patients.

Methods: Fifty-one patients with CFLD were followed for 2 years. Adjacent to biochemical and biannual examination, transient elastography was performed in conjuction with ultrasound evaluation. Williams ultrasound scoring system was used for grading the liver alterations in stage 1 – mild (points <3), stage 2 – moderate (4–8 points), stage 3 – severe (>8–9 points). Control group age matched included 36 healthy children.

Results: Outcomes at baseline showed that most of patients 62.74% (32 patients) had moderate stage 2 liver disease, with median value liver stiffness 8.8 kPa. Only 9.8% (5 patients) had liver cirrhosis, with ultrasound score >9 and median elasticity 14.6 kPa. 14 patients (27.4%) had CFLD stage 1, ultrasound features consisting in hyperechogenity and mean elasticity was increase 7.6 kPa compared to controls, suggesting fibrosis in cases considered as liver steatosis. Median fibrosis score in children with CFLD was 10.33 kPa compared to controls.

Conclusion: Elastography is a valuable method for evaluation of liver fibrosis in CF patients, helping with differentiation between steatosis and fibrosis. Early CFLD diagnosis with consequent intervention could prolong CF children's life.

219 Prevalence of cystic fibrosis related liver disease in Northern Ireland

S.A. Rowan[1,2], I. Moore[2], W.J. Cash[3], D. Downey[1,2], J.S. Elborn[1,2], J. Rendall[1,2]. [1]*Queen's University Belfast, Centre for Infection and Immunity, Belfast, United Kingdom;* [2]*Belfast Cystic Fibrosis Centre, Belfast, United Kingdom;* [3]*Royal Victoria Hospital, Liver Unit, Belfast, United Kingdom*

Introduction: Cystic fibrosis related liver disease (CFRLD) reported incidence is between 27% and 35%. A spectrum of hepatobiliary disease can occur in cystic fibrosis (CF), with a small number progressing to liver failure. Liver cirrhosis is the largest non-pulmonary cause of death occurring in approximately 2.5% of CF mortality.

Objective: To assess the prevalence of CFRLD in the N. Ireland adult CF population.

Methods: A retrospective review of case records of the CF adult population was undertaken. Age, FEV1, BMI, microbiology status, USS and OGD results, documented gastrointestinal and hepatic disease were recorded. Evidence of referral to a gastroenterologist/hepatologist and liver transplant status were also recorded.

Results: 241 patients were identified (135M/106F). Mean (SD) age 31(11.5), FEV1% 71.4(22.4), BMI 23(3.6) kg/m^2. 45% were infected with *P. aeruginosa*, 9% BCC and 38% non-*Pseudomonas*. The commonest reported genetic mutations were 34% F508/F508, 10% F508/R117H, 5% F508/G551D and 25% F508/other. 85% had an USS, with splenomegaly present in 10%, fatty liver in 16% cirrhosis in 4%. 11% had evidence of biliary disease and 8% had a final diagnosis of CFRLD. In the CFRLD group 60% were F508/F508. CFRLD is more common in F508 homozygotes ($p = 0.01$). One death from liver disease occurred and one is transplant listed.

Conclusion: This is a lower than expected prevalence of liver disease in comparison with data from Nash et al 2008 (cirrhosis in 28%, fatty liver in 3% and splenomegaly in 6%). This may be due to inaccurate coding or potential genetic differences seen in N. Ireland. F508 homozygotes appear to have a higher incidence of CFRLD than other mutations.